Podiatric Dermatology as a Manifestation of Systemic Disease
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Case #1

The Facts
• Middle-aged male
• Symmetric, painful > pruritic nodules of the plantar surfaces
• Associated with systemic vasculitis, hypereosinophilia, asthma, and allergic rhinitis
• Antibodies to p-ANCA detected

Case 1 Answer?
A. Leukocytoclastic vasculitis
B. Wegener’s granulomatosis
C. Subcutaneous granuloma annulare
D. Churg-Strauss Syndrome
E. Polyarteritis nodosa

D. Churg-Strauss Syndrome
Churg-Strauss Syndrome
• Syndrome of Vasculitis, eosinophilia, and asthma/allergic rhinitis
• Variable course, but usually responds well to corticosteroids
• May be associated with various other, more serious autoimmune disorders

Case #2

The Facts
• Young adult male
• Symmetric, painful papules peripherally lining the plantar surfaces
• Associated with longitudinal striations of the nails
• Father had a similar disorder
• Biopsy showed acantholysis and intracorneal hemorrhage
Case 2 Answer?

A. Punctate keratoderma  
B. Porokeratosis punctata  
C. Acral Darier’s disease  
D. Erythroderma Verruciformis  
E. Ectodermal dysplasia

C. Darier’s Dss;  
Acral Hemorrhagic Variant

Darier’s Dss;  
Acral Hemorrhagic Variant

• Autosomal dominant genodermatosis  
• May be associated with, and masked by tinea  
• A fundamental defect in the synthesis of tonofilament-desmosome complexes  
• Chromosomal defect mapped to 12q23-24.1

Case #3

The Facts

• Elderly female  
• Large tense bullae affecting right hallux, eventuating in sloughing of skin  
• Punch biopsy showed sub-epidermal clefing, and numerous eosinophila  
• Immunofluorescence showed linear reactivity at the dermal-epidermal junction  
• Development of vesicles on trunk and upper extremity over following weeks
Similar Cases
Case 3 Answer?
A. Diabetic bullous dermapathy  
B. Bullous pemphigoid  
C. Porphyria  
D. Pemphigus vulgaris  
E. Epidermolysis bullosa simplex

B. Bullous Pemphigoid

- The most common subepidermal bullous disease
- Usually a disease of elderly men
- Characteristic urticarial lesions precede tense subepidermal bullae
- May be a medical emergency; lethal if untreated
- Caused by auto-antibodies directed against proteins in hemidesmosomes

Case #4

Similar Cases
The Facts

- Upper middle aged male
- Warty papules beginning on forefeet
  - Slowly progressive over years
  - No tendency toward resolution
  - Non-painful, not pruritic
- Venous incompetence with longstanding nonpitting venous stasis
- Type 2 diabetes mellitus
- No travel history

Case 4 Answer?

A. Vascular malformation diffusum
B. Filariasis
C. Elephantiasis nostra verrucosa
D. Stewart-Treves syndrome (angiosarcoma in stasis)
E. Diffuse lymphangiomas
C. Elephantiasis nostra verrucosa

Elephantiasis Nostra Verrucosa
- Warty papules that result as an end-stage phenomenon secondary to chronic lymphedema/hypoxemia
- Essentially an end-stage scarring process
- May be slowed, but not reversed

Case #5

Case 5 Answer?
A. Pseudo-porphyria
B. Cirrhosis-related stasis bullae
C. Pemphigus vulgaris
D. Epidermolysis bullosa simplex
E. Acquired epidermolysis bullosa

A. Pseudo-Porphyria

The Facts
- 45 year old woman
- Recent urinary tract infection treated with sulfonamides
- Worsens with sun exposure
- Urine and fecal porphyrins at normal levels
- Biopsy revealed subepidermal clefting without inflammation
- Negative immunofluorescence
Pseudo-Porphyria

- Pseudoporphyria exhibits clinical findings that parallel porphyria, but with normal porphyrin levels
- Most often drug induced with antibiotics and NSAIDs common culprits
- Sun-exposed surfaces, esp. the dorsal hands and feet
- Resolves upon removal of offending agent

Case #6

The Facts

- 17 year-old male
- Tennis player
- Life-long occurrence of blisters after rigorous activity
- Biopsy demonstrates subepidermal clefing without inflammation
- Negative immunofluorescence
- Recently diagnosed as “pre-diabetic”
- No drug history; normal porphyrin levels
D. Epidermolysis Bullosa Simplex (Weber-Cockayne type)

Epidermolysis bullosa simplex
- Called “Epidermolytic” bullosa
  - As opposed to “junctional” or “dermolytic”
  - Mechanobullous disorders
- Congenital defect in the production of keratins 5 and 14 (incidence 1:50,000)
  - Koebner variant (generalized)
  - Weber-Cockayne variant (localized)
- Supra-basilar blisters (non-scarring)
- Palliative treatment

Similar Case

Case #7
The Facts

- 53 year old woman
- Penicillin prescribed for an infected sacral ulceration
- Type 1 diabetes mellitus
- No history of sun exposure
- Biopsy revealed intraepidermal clefting with minimal chronic inflammation
- Negative immunofluorescence

Diabetic Bullous Dermopathy

- Subepidermal blisters with minimal associated inflammation
- Negative immunofluorescence
- Usually a relatively localized process
- Etiology not clear
- Treatment is largely palliative

Case 7 Answer?

A. Cirrhosis-related stasis bullae
B. Linear IgA dermapathy of diabetes
C. Pemphigus vulgaris
D. Diabetic bullous dermapathy
E. Acquired epidermolysis bullosa

D. Diabetic Bullous Dermopathy
The Facts
- Longstanding type 2 diabetic
- Daily aspirin therapy for atrial fibrillation
- Slowly evolving brown macules coalescing to form patches
- Some associated skin atrophy
- Lesions do not appear to resolve

Case 8 Answer?
A. Necrobiosis lipoidicum
B. Schamberg’s disease
C. Stasis hemosiderosis
D. Coumadin-related purpura
E. Diabetic pigmented pretibial patches

Diabetic Pigmented Pretibial Patches
- Seen in longstanding diabetics
- Progressive atrophic pigmented macules and patches of the anterior leg
- Related to microvascular compromise

Case #9
The Facts

• 23 year old well controlled type 1 diabetic
• Slowly expanding pink-yellow plaques/nodules on lower legs
• Have ulcerated, but healed on their own
• Non-painful, non-pruritic
• No exasperating associations

Similar case
Case 9 Answer?
A. Nummular dermatitis
B. Necrobiosis lipoidica
C. Tinea corporis
D. Granuloma annulare
E. Erythema nodosum

B. Necrobiosis Lipoidica (diabeticorum)

Necrobiosis Lipoidica (diabeticorum)
- Disorder of collagen degeneration
- Thought to be related to microangiopathy
- 50% of cases arise in association with diabetes
- Slowly progressive plaques and nodules which may ulcerate
- Pathogenesis unclear; may be related to granuloma annulare

Case #10

The Facts
- Middle aged woman
- History of severe alcohol abuse and cirrhosis
- Glucose tolerance normal
- Biopsy showed subepidermal blisters with no inflammation
- Immuno fluorescence negative
- Blistering worsened with ultra-violet light exposure

Case 10 Answer?
A. Cirrhosis-related stasis bullae
B. Porphyria
C. Pseudoporphyria
D. Pemphigus vulgaris
E. Epidermolysis bullosa simplex
B. Porphyria Cutanea Tarda

**Porphyria Cutanea Tarda**

- The most common form of porphyria
- Caused by a defect in the biosynthesis of heme leading to excess production of porphyrin precursors
- Related to defects within the liver (others related to defects within RBCs)
- Usually related to ETOH, cirrhosis, or estrogen
- Increased uroporphyrins in urine and plasma, and coproporphyrins in feces
The Facts

- Upper middle-aged female
- Recurrent tissue breakdown on the dorsum of feet
- Cultures showed pseudomonas
- Would clear up when hospitalized on IV antibiotics, only to recur following release

- Nondiabetic
- 3+ pitting edema

Case 11 Answer?

A. Pseudomonas carrier state
B. Acro-angiodermatitis
C. Common stasis ulcers
D. Erosion superscriptum
E. Candida interrigo

B. Acro-angiodermatitis (of Mohly)

Acro-angiodermatitis (of Mohly)

- An alternate expression of end-stage stasis/hypoxemia
- An intra-dermal proliferation of atypical vessels, once designated as 'PSEUDO-KAPOSI'S SARCOMA'
- May be slowed but not reversed
Similar Cases
The Facts

- Middle-aged females
- Slowly progressive papules and plaques over several months
- Intensely pruritic
- Recently diagnosed Hepatitis C infection
- OTC corticosteroids of little help
- No significant family history

Case 12 Answer?

A. Hypertrophic lichen planus
B. Acral psoriasis
C. Pityriasis rubra pilaris
D. Paraneoplastic keratoderma
E. Palmoplantar keratoderma
A. Acral Lichen Planus, Hypertrophic type

- Men 20-60 years of age
- Women incidence increases with age
- May be associated with Hep C, some drugs (gold), or HIV
- 2/3 of patients have active disease for less than 1 year
  - Hypertrophic / acral lichen planus may last for decades
- First line therapy is corticosteroids +/- non-steroidals

Case #13

Similar Cases
The Facts
- 48 y/o woman with slowly progressive acral plaques
- Mildly pruritic
- Fine scales over an erythematous base
- HIV negative
- No significant family history
- OTC corticosteroids of no value

Case 13 Answer?
A. Non-hypertrophic lichen planus
B. Acral psoriasis
C. Pityriasis rubra pilaris
D. Paraneoplastic keratoderma
E. Palmoplantar keratoderma
B. Acral Psoriasis

- The most debilitating and recalcitrant form of psoriasis
- 80% of cases are limited to acral surfaces
- M = F
- Polygenic trait
- Two peaks: 22 y/o and 55 y/o
  - Earlier onset coincides with severe expression
  - Commonly triggered
    - Trauma, stress, ETOH, HIV, drugs (Li, Gliubocort)

Case #14

Similar Cases
The Facts

- Lower middle-aged woman
- Bilateral painful ulcers which evolve into painless scars at the inner ankles
- History of lupus erythematosus
- Minimal stasis
- No peripheral ischemia
- No history of sickle cell trait/disease

Case 14 Answer?

A. Sickle cell disease
B. Leukocytoclastic (allergic) vasculitis
C. Polyarteritis nodosa
D. Livedoid vasculitis
E. Occult stasis ulcerations

D. Livedoid Vasculitis (atrophy blanche)

Livedoid vasculitis (atrophy blanche)

- A disease of young and middle-aged women
- Manifests as painful sites of erythema with ulceration, evolving scar formation
- Medial lower leg and ankles
- Fundamentally a vaso-occlusive disorder affecting arteries and veins
- May be primary (idiopathic) or secondary
  - Connective tissue Dss, venous incompetency

Case #15
The Facts
- 43 year old woman with rapidly arising painful macules on the skin of both lower extremities
  - Associated with some / minimal pain
- Some macules disclosed central ulcerations
- Transient low-grade fever
- Biopsy showed a small to intermediate-sized vessel necrotizing vasculitis
  - Neutrophils and histiocytes
- C-ANCA positive

Case 15 Answer?
A. Rheumatoid vasculitis
B. Leukocytoclastic (allergic) vasculitis
C. Polyarteritis nodosa
D. Wegener’s granulomatosis
E. Superficial thrombophlebitis

Wegener’s Granulomatosis
- A systemic form of vasculitis presenting in the skin in 14% of cases
  - When the skin is involved it is often the presenting complaint
- Uniformly fatal if untreated due to lung and kidney involvement
- Associated with the presence of C-ANCA
- Usually responds well to corticosteroids and methotrexate
The Facts
• 18 year old girl
• Arthritis, abdominal pain, and hematuria
• Hemorrhagic vesicobullae
• Biopsy revealed a superficial vasculitis
• Immunofluorescence demonstrates IgA immunocomplexes around vessels

Case 16 Answer?
A. Rheumatoid vasculitis
B. Leukocytoclastic (allergic) vasculitis
C. Linear IgA immunobullous Dss
D. Superficial thrombophlebitis
E. Henoch Scholein Purpura
Henoch Scholein Purpura
- Purpuric rash with the triad of
  - Arthritis
  - Abdominal pain
  - Hematuria
- IgA Immune complex deposition within vessel wall
- Common renal involvement in adults
- Self limited in most cases

Case #17

Similar Cases
The Facts

- 56 year-old man
- Seasonal onset (winter) of purpuric macules involving digits
  - Painful
  - Resolve in 1-2 months in most cases
- Works in chicken processing plant in WA
- No significant medical history
- Normal cold precipitation test
Case 17 Answer?
A. Cryoglobulinemia  
B. Rheumatoid vasculitis  
C. Leukocytoclastic (allergic) vasculitis  
D. Perniosis  
E. Superficial thrombophlebitis

D. Perniosis (chilblains)

Perniosis (chilblains)
• A form of low-grade lymphocytic vasculitis
• Induced by cold exposure in most, but not all, cases
• Exacerbated during the winter months
• Once thought to be an expression of lupus erythematosus
• Treated with warmth and low dose nifedipine

Case #18

The Facts
• 52 year-old male
• Works as a gardner
• Extremely proliferative wound with rolled edges
• Remote history of irritable bowel syndrome
• Positive pathergy test
Case 18 Answer?

A. Pyoderma gangrenosum
B. Basal cell carcinoma
C. Deep fungal infection
D. Squamous cell carcinoma
E. Stasis ulceration

Pyoderma Gangrenosum

• A disorder leading to florid and exaggerated responses to injury
• Pathergy test
• May be associated with ulcerative colitis, rheumatoid arthritis, paraproteinemias, others
• Usually responds best to oral corticosteroid therapy

Case #19

The Facts
• 37 year old woman with a history of "irritable bowel syndrome"
• Inflamed nodules on the lower legs, exquisitely tender
• Has requested to have nodules excised due to pain
• Recent discovery of "abnormal" chest radiograph

Similar case
Case 19 Answer?
A. Polyarteritis nodosum
B. Granuloma induratum
C. Erythema nodosum
D. Lupus profundus
E. Pancreatic panniculitis

C. Erythema Nodosum
A form of granulomatous panniculitis marked by extremely tender nodules over the skin of the lower legs
- Less often upper extremity
- Often seen in association with Crohn’s Dss, sarcoidosis, or histoplasma exposure
- Best treated with oral anti-inflammatory therapy

Case #20

The Facts
- Patient with dystrophic nail units bilaterally
- Tinea pedis bilaterally
- Fungal infection not confirmed with histopathology
- Placed on oral Lamisil continuous dosing
- Soon began developing skin lesions
- Positive ANA, anti-Ro (SS-A) antibodies were detected

Case 20 Answer?
A. Subacute Lupus erythematosus
B. Polyarteritis nodosum
C. Id reaction
D. Viral exanthem
E. Cutaneous drug eruption

A. Subacute Lupus Erythematosus
Subacute Lupus Erythematosus

- Occurs in genetically predisposed individuals
  - HLA-B8, HLA-DR3, etc.
- Females over males (4:1); 40-60 years of age
- A strong association with anti-Ro (SS-A) autoantibodies
- May be seen in association with SLE, Sjogren syndrome, Compliment C1d deficiency, or may be drug induced

Case #21

The Facts

- 73 year old woman
- Referred to podiatrist by a central Florida internist for I&D of a carbuncle
- Erythematous nodule present for roughly 1-2 months
- Biopsy performed

Diffuse Large B-cell Lymphoma

The Facts

- Subsequent formation of leg nodules on ipsilateral leg
  - Coincided with the onset of fever and malaise
Case 21 Answer?
1. Toxic arthropod bite reactions
2. Metastatic melanoma
3. Diffuse large B-cell lymphoma (of the leg)
4. Deep fungal infection
5. Erythema nodosum

C. Diffuse Large B-cell Lymphoma (of the leg)

Diffuse Large B-cell Lymphoma
- High-grade lymphoma
- Poor prognosis with overall 5 yr survival of about 50%
- Treatment
  - Solitary lesions treated with excision or external beam radiotherapy
  - Multiple lesions treated with radiotherapy
  - Systemic involvement treated with chemotherapy

Case #22

Similar Cases
The Facts
• 53 year-old Caucasian woman
• History of multiple myeloma, with associated immunosuppression and end stage renal dis
• Elevated plasma lipids secondary to corticosteroid therapy
• Currently maintained on hemodialysis

Case 22 Answer?
A. Eruptive molluscum contagiosum
B. Diffuse superficial actinic porokeratosis
C. Acquired perforating dermatosis
D. Eruptive xanthoma
E. Guttate psoriasis

C. Acquired Perforating Dermatosis (Kyrle Dss)

Acquired Perforating Dermatosis(Kyrle Dss)
• Poorly understood disorder consisting of multiple pruritic cyst-like lesions
• In this context designated as Perforating Disease of Chronic Renal Failure
• Associated with:
  – Diabetes mellitus
  – Renal Dis
  – Hepatic disease
  – Congestive heart failure

Case #23

The Facts
• 27 year old female
• Type 1 diabetes mellitus, recently becoming insulin resistant
• Rapidly arising papules over the heel and arch bilaterally
• Papules are firm and painless
Case 23 Answer?
A. Calcinosis cutis
B. Epidermal inclusion cysts
C. Foreign body implantation
D. Xanthoma diabeticorum
E. Granuloma annulare

D. Xanthoma Diabeticorum
- A form of eruptive xanthoma which arise rapidly and may be widespread
- Associated with elevated triglycerides/chylomicrons, and to a lesser extent, cholesterol
- May precipitate acute pancreatitis

Case #24

Similar Case
Case 24 Answer?
A. Verrucous xanthoma
B. Elephantiasis nostras verrucosa
C. Mosaic warts (HPV serotype 2)
D. Verrucous carcinoma
E. Mosaic warts (HPV serotype 4)

Case 25 Answer?
A. Tinea nigra
B. Acral acanthosis nigricans
C. Psoriasis
D. Minocycline pigmentation
E. Schoenberg's disease

Case #25

The Facts
- 52 year-old African-American male
- Non-pruritic velvety areas of dark pigmentation over plantar and volar surfaces
- No history of diabetes or obesity
- Recent onset of deep-seated nonproductive cough
- X-ray shows dense LLL infiltrate
Acral Acanthosis Nigricans

- Progressive pigmentation of the acral surfaces with altered skin texture
- Originally described as a paraneoplastic condition
  - More recently hypothesized simply to be reactive secondary to rubbing or scratching

Thanks!